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PROCEEDINGS

OF THE

PATHOLOGICAL SOCIETY

OF

PHILADELPHIA

New Series, Vol. IX. No. 2, 1906

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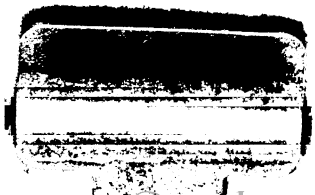

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A Study of the Elastic Tissues in the Parous and the Non-parous Uterus.

BY BROOKE M. ANSPACH, M.D.

IN order to make a description of the elastica plain, it will be necessary to state that according to Kreitzer there are four layers of muscle in the uterine wall. There is first the layer containing the large bloodvessels, spoken of as the stratum vasculare, which occupies the largest part of the myometrium; running between this and the mucosa is a layer whose general direction is longitudinal, spoken of as the stratum submucosum. An outer layer, running longitudinally directly under the serous coat is given the name stratum subserosum. The irregular fibers between the later stratum and the vascular layer are known as the stratum supravasculare.

The elastic tissue of the uterus is derived from two main sources. First, the serous coat; second, the bloodvessels. In the subserous and in the supravascular layers the elastic fibers are very rich. They surround the individual muscle cells in the outer layer, forming elastic perimysia. They are not subdivided to this extent in the supravascular layer. Their general direction is toward the endometrium and as the outer muscular layer is longitudinal and the supra-

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vascular layer is more or less circular, they run at right angles to these muscle bundles. In the vascular layer the elastica is mostly confined to the vascular channels. In the submucosa there is practically no elastica except in the vessels. The elastic tissue of the subserous and supravascular layers appears as fine, unbroken fibers of a more or less uniform thickness. Many of them are quite straight, especially those taking a centripetal course. In the vascular layer elastic tissue is found surrounding the lymph spaces, the capillaries, the veins, and the arteries. About lymph spaces they appear as short, wavy fibers or as black points; their distribution is quite irregular and the number is always small. This is true also of the capillaries. In the veins there is a collection of elastic tissue between the inner and the outer coat. It is quite irregular and forms no distinct limiting band, as in the arteries. In the small and medium sized arteries there is a distinct limiting membrane of elastic tissue between the intima and the media, the internal elastic membrane; and between the media and the adventitia in the larger vessels is another, the external elastic membrane.

Pick draws attention to the arrangement of elastic tissue in the uterus as being highly significant. The preponderance of the fibrils in the subserous and the supravascular layers, their definite direction (centripetal) toward the endometrial cavity, their fine subdivision so that in the outer layer they surround each individual muscle cell, all of this he believes shows they have a very well-defined purpose. They serve not only for the support of the blood and lymph capillaries and the nerves, but further than this they reinforce the muscular action of the uterus, protect it from being overstretched, and make easier its return to a passive state after either distention or contraction.

The study I have made is based on twenty-three specimens. Five of these were uteri from women who had uncontrollable metrorrhagia to which none of the usual causes could be ascribed. The other eighteen were specimens removed along with the appendages in pelvic inflammatory disease. The uterus in such cases is not much affected.

The subjects from which these uteri were taken varied in age from seventeen to fifty-six. Five of the women were nulliparous,

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thirteen were parous. The five uteri from the metrorrhagia cases were all multiparous.

Sections were taken from the same part of the uterus in each case; they were cut of the same thickness and they were stained for the same length of time by Weigert's resorcin fuchsin method.

There are very striking changes in the elastic tissue as the result of pregnancy, and the greater the number of pregnancies the greater are these changes. It is quite easy to say from a section of the muscle of the uterus stained by the resorcin fuchsin stain whether the woman has had children or not. The alterations of the elastica affect not only that in the two outer layers of the myometrium, but also that in the vascular channels. The elastic tissue in the subserous and in the supravascular coat becomes increased in amount and the fibers are shorter, thicker, show a tendency to become clumped together, and are curly rather than straight.

In the vessels wherever elastic tissue is present normally the amount is very much increased; the fibers showing the same tendency to become broken, thick and gathered into clumps. The vessel changes, which were observed in these cases may be spoken of as intramural and perivascular. The intramural changes in the arteries consist of a broadening and splitting of the internal elastic membrane. It no longer consists of a fine single line, but is irregular and three or four times its normal thickness. The elastica of the veins in this form (intramural) is similarly increased.

Another form of elastic hypertrophy, the perivascular, occurs in the adventitia. The entire adventitia is represented by heavy masses of elastic tissue in which no small fibrils are seen—the tissue appears a dense black. In sections stained by Van Gieson's stain these same areas appear bright yellow, and with hematoxylin and eosin they are granular, pink and contain scattered well-preserved nuclei. The internal elastic membrane in such cases may be fairly normal. This degeneration, perivascular, increases in some cases until it occupies the entire wall of the artery. Such a vessel is bright yellow by Van Gieson's stain and of a granular pink color by eosin hematoxylin, while densely black with Weigert's stain.

The same dense deposit of elastic tissue occurs in the outer coat of the larger veins.

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These areas here appear bright yellow by Van Gieson's stain and pinkish yellow by hematoxylin eosin.

From this study it appears that the above alterations in the elastic tissue are the result of pregnancy. They are not the result of age, for in two nullipara aged thirty-seven, they did not occur and in a primipara aged forty-one they were not marked.

These changes in the uterine vessels which are often spoken of as arteriosclerosis are the result of pregnancy and must be considered normal. They are much more constant and notable than the increase in the relative proportion of fibrous tissue which often follows pregnancy. *December* 14, 1905.

Congenital Malformations of the Heart; A Series of Cases.

By ALLER G. ELLIS, B.Sc., M.D.

(From the Laboratories of the Jefferson Medical College and the Philadelphia General Hospital.)

CONGENITAL malformations of the heart, both singly and in their varied combinations, form a group of cases of perennial interest alike to the clinician and to the pathologist. The characters making up the symptom-complex during life, although not constant and hence rarely leading to accurate diagnosis of the exact lesion, are nevertheless quite well known; the possibilities of autopsy findings are bewildering. A study of over 600 reported cases, which by no means exhausts the enormous literature on the subject reveals forty-five different single or combined lesions of the heart and great vessels. This fact renders impracticable my original intention to classify the cases according to the nature of the malformation, and I shall speak in particular only of those lesions, seven in number, which are represented by the specimens on exhibition. It is interesting to note, however, that the forty-five cases of cardiac malformation found among the 3875 autopsies, with complete records, performed at the Philadelphia Hospital during the past thirteen years are grouped as follows:

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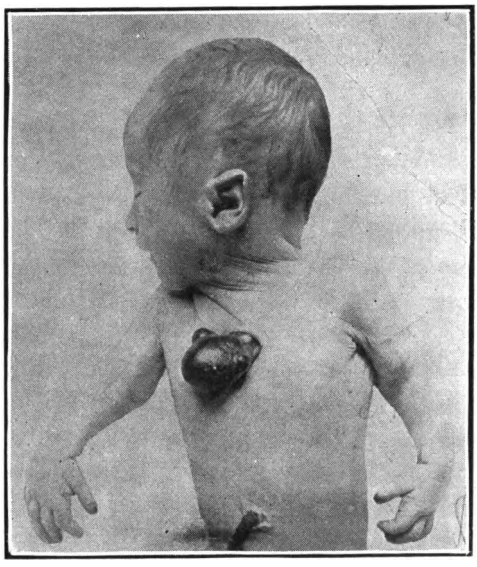
Patent foramen ovale	31
Patent ductus arteriosus	1
Absence of auricular septum	1
Absence of anterior portion of auricular septum	1
Patent foramen ovale and patent ductus arteriosus	10
Patent foramen ovale, patent ductus, pulmonary stenosis and perforate interventricular septum	1
Patent foramen ovale, patent ductus, and transposition of aorta and pulmonary artery	1

In the sixty autopsies I have held elsewhere than at the Philadelphia Hospital I have found five instances of patent foramen ovale.

CASE I.—*Ectopia Cordis.* This specimen was recently acquired by the museum of the Jefferson Medical College through the courtesy of Dr. George Cunningham. The child is a full-term male.



Case I.

The lateral halves of the sternum are united except for a distance of 2 cm. at the lower extremity, the gap between the parts being 5 cm. wide. The heart is entirely external to the thoracic wall, the vessels passing in through an oblong and diagonally placed opening 2 by 3.5 cm. in size. The organ is covered by visceral pericar-

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dium only and can be drawn some distance from the chest by exerting traction upon the great vessels. The latter structures, as well as the heart, are entirely free from the margin of the opening in the skin and underlying tissues. The auricles are rounded and project prominently; the apex is blunt and is formed nearly equally by the two ventricles. The heart is essentially normal in size. As observed by the nurse, the organ performed its functions properly while the child lived, a period of twenty-three and one-half hours.

Ectopia of the heart may be associated with fissure of the sternum, exocardia or pectoral heart, as in the present instance, or with an intact sternum the organ may be situated in the neck (cervical heart) or in the abdomen (abdominal heart). Instances of the first-named group are the most common and include those with entire separation of the sternum and those with superior and inferior defect, respectively. The condition is relatively rarely found alone, being usually accompanied by protrusion of the abdominal viscera. Peacock[1] cites thirteen cases of the various types reported by others and describes another specimen in the Museum of St. Thomas' Hospital. Ballantyne[2] mentions nine additional cases, and states that others have been reported. He refers to nine cases of the sternoepigastric type collected by Taruffi,[3] but as the entire sternum and the abdominal wall to the umbilicus were divided, Ballantyne regards these cases as more properly belonging under gastroschisis. I have obtained references to three cases not included in the series mentioned.

When the halves of the sternum are entirely separated, the pericardium is usually absent; the heart itself may be normal or exhibit varied malformations. In a case of this type reported by Grant[4] the subject was a seven months' fetus, which lived six hours in an incubator. The halves of the sternum were 5 cm. apart at the upper and 10 cm. at the lower end, and the extreme upper portions of the recti muscles were slightly separated. The visceral pericardium was reflected from the heart and joined to the skin. At each systole the heart was tilted upward, the apex reaching the chin.

An example of superior defect of the sternum is the case of Sidney Jones, cited by Peacock. It differed from my case in that the skin was prolonged over the great vessels and thence to the base of the

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heart, the epithelium extending a short distance on the auricular walls. The child was born at the eighth month and lived thirteen hours. When the opening involves the lower part of the sternum and the epigastric region the pericardium is not unusually present and the person may reach adult life. In the case reported by Goode[5] the manubrium was intact, the heart protruding below through the passage of the bloodvessels, with the covering of which the skin at the lower border of the foramen was continuous. The heart was normal in size and shape; it was kept bathed with olive oil and covered by a pasteboard cone. The child lived sixteen days. In the absence of defect in the sternum an opening in the diaphragm may allow of the condition of abdominal heart, with or without external tumor. In the case reported by Holt,[6] a living child five months of age, with umbilical hernia, the heart was situated below the ensiform cartilage, the circulation beng fairly well maintained. Peacock cites the case of Deschamps who, at autopsy upon the body of a soldier, found the heart occupying the usual position of the right kidney, the vessels passing into the thorax through an opening in the diaphragm.

Ballantyne states that ectopia cordis is rare in human beings and presents several difficult problems in teratogenesis; the theory of arrested development is permissible. Non-separation of the membranes may prevent fusion of the body walls in the middle line and amniotic adhesions may hold the heart forward. In cases in which the heart is covered by skin, the pressure and adhesions may have ceased early enough to allow the walls to close imperfectly. As the presence of the heart external to the body is incompatible with the maintenance of life, an interesting question is raised by the case of Lannelongue. In that instance a thin membrane which covered the heart sloughed on the fifteenth day; the skin margins were then united over the heart and the patient recovered. It appears reasonable that when the conditions are even in the least degree favorable to surgical interference, such a step should be carefully considered as a possible means of saving life.

CASE II.—*Transposition of the Aorta and Pulmonary Artery; Patent Foramen Ovale; Patent Ductus Arteriosus.* This case has elsewhere[7] been reported at length and will here be only briefly described.

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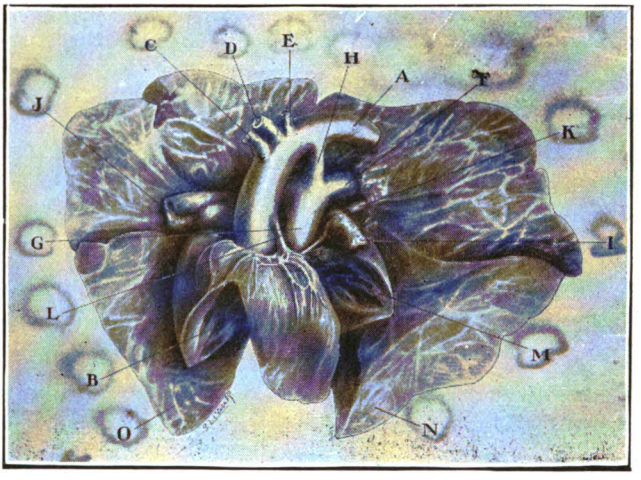
The subject was a male infant who died at the age of thirty-four days. At birth it weighed seven pounds and the day before death seven and one-quarter pounds. Respiratory difficulty was at all times present and cyanosis was marked during the first few days. Later the blueness became less prominent, especially at short periods, during which it almost disappeared. The child died during an attack of labored and irregular breathing accompanied by extreme cyanosis. The pathologic diagnosis was: transposition of the aorta and pulmonary artery; patent foramen ovale; patent ductus arteriosus; hypertrophy of right ventricle; partial atelectasis of left



Case II.

lung; general visceral congestion. The heart is essentially normal in size. In thickness the wall of the right ventricle is 7 mm., of the left, 5 mm. Neither the aorta nor pulmonary artery shows evidence of stenosis. The foramen ovale is patulous in the shape of a slit-like opening 0.5 cm. long. The ductus arteriosus allows with ease the passage of a probe 2 mm. in diameter.

Cases of transposition of the great vessels of the heart are not extremely rare, probably as many as seventy-five being recorded. The malformation in origin is developmental rather than fetal in point of time, being due to lack of rotation of the septum in the

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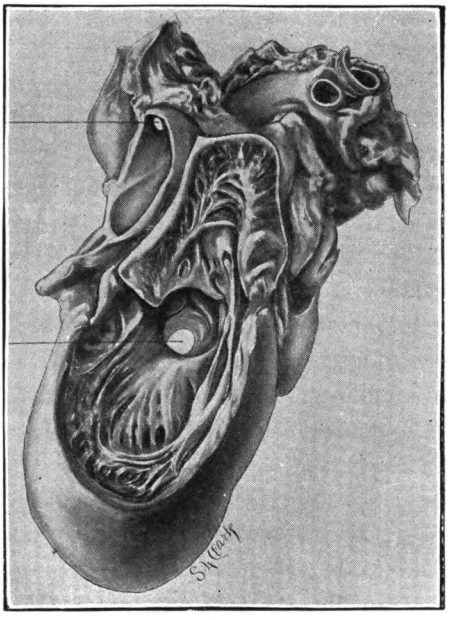
aortic bulb. It is incompatible with long life, practically all of the subjects dying before the age of three years. A coexistent opening in the interventricular septum favors the prolongation of life. There are no pathognomonic physical signs or symptoms of transposition and the clinical diagnosis can be made only by inference; even this is rarely done.

CASE III.—*Patent Interventricular Septum; Pulmonary Stenosis; Patulous Foramen Ovale; Patulous Ductus Arteriosus.* This speci-



Case III.

men was obtained at an autopsy held by Dr. Coplin at the Philadelphia Hospital upon a male child of eleven months, the diagnosis of the three first-named lesions having been made during life by the attending physician, Dr. McKee. The body was extremely emaciated and the gums were spongy. The cavities of the right side of the heart, which is a globular organ, were found distended with blood. The right auricle communicates with the left through an oblique slit in the foramen ovale 3 mm. in maximum size. The pulmonary

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artery arises from the right ventricle at about its normal position, the conus pulmonalis being narrowed. At its point of origin the artery is 7 mm., and 1 cm. beyond this, 2 cm. in circumference and the orifice is guarded by a single, very thick, crescentic leaflet. From the artery 2 cm. beyond its origin springs the ductus arteriosus, which enters the aorta 2.5 cm. from its valves. The right ventricle at the base communicates with the left through a somewhat triangular opening in the membranous septum, the apex being directed toward the apex of the heart, and each side measuring 4 mm. in length. As a result of this septal defect both ventricles open into the aorta immediately over the septum. The wall of the right ventricle is 7 mm., of the left, 5 mm. in thickness.

Of this combination of lesions perforate interventricular septum is not infrequent, though not at all uncommon. The opening is usually anterior or posterior, most often the former, to the pars membranacea, or undefended space. Hart[8] states that perforation of the membranous septum can occur only as the result of inflammation and is very rare. The fact that deficiency is usually in the posterior part of the anterior segment of the septum is of interest in connection with the question of heart block as related to the auriculoventricular muscle bundle of His. The latter structure passes down along the upper edge of the muscular septum where that tissue joins with the membranous portion and terminates below the root of the aorta. The possibility of interference with this band when the defect is in the posterior segment of the septum appears worthy of note, although I know of no recorded case of heart block associated with such malformation. The presence of deficiency in any one of the three parts of the septum is not incompatible with the attainment of adult life. Cyanosis may or may not be a symptom of the lesion. Reiss has called attention to the fact that cyanosis may be absent in early life and later make its appearance.

Communication between the ventricles is due to maldevelopment of one of the several component parts of the septum and not uncommonly occurs in connection with stenosis or atresia of the pulmonary artery. Keith[9] furnishes an admirable discussion of this question in connection with a series of 30 malformed hearts, which he exhibited. Of the 185 specimens of malformation of the heart in the London

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museums, all of which he has studied, 113 show developmental error in the conus pulmonalis and 22 additional organs congenital stenosis of the pulmonary valves alone. In a large number of the 113 specimens there is an interventricular opening. Keith directs attention to three features in the septal wall of the infundibular portion of the right ventricle which are at present little noticed by anatomists. They consist of a furrow and two muscle bands and their evolution, together with the expansion of the conus pulmonalis, has much to do with the closure of the interventricular foramen. The persistence of the foramen is usually accompanied by patulous foramen ovale. Antemortem diagnosis of ventricular communication is possible, as mentioned in the present instance. Most prominent is a murmur which is loud, systolic or almost continuous in time, and may be heard over the larger part of the chest; it is loudest along the course of the interventricular septum.

Pulmonary stenosis is considered by the great majority of writers to be the most common of congenital malformations of the heart; this is well attested by the statistics of Keith and by Peacock's 119 cases among 181 of congenital anomaly. Complete atresia of the artery is less common than is stenosis. Stifel states that of the cases of atresia the artery is only a string-like cord in 13 per cent., the ductus arteriosus is open in 82 per cent., the foramen ovale in 61 per cent., and there is a defect in the interventricular septum in 76 per cent. In simple stenosis the ductus is seldom, the foramen ovale frequently, open. In one of McCrae's[10] cases of atresia with deficient auricular septum and open ductus a fetal systolic murmur was demonstrated to students before the birth of the child. In this subject there was also transposition of the viscera, a condition of which Arneill in 1902 collated more than 300 cases. In some of these instances the malposed heart is also malformed, but the former condition does not appear specially to predispose to malformation. A moot point regarding pulmonary stenosis is its influence upon the incidence of tuberculosis of the lungs. The common belief is that this malformation does favor the disease in question. Burke[11] regards this occurrence of pulmonary tuberculosis, which was present in all three of his cases, as due not to pulmonary stenosis but to coexisting narrowing of the aorta.

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Persistence of the ductus arteriosus is a postnatal malformation. It is exceedingly rare as a single lesion, being almost always associated with one or more other cardiac or vascular defects. Clinically, an interesting point is the possibility of diagnosis. An elaborate symptom-complex of the lesion is given by various writers, but the most prominent feature regarding them is the regularity with which they fail. Arnheim[12] reports a case in which what seemed a clear diagnosis of open ductus was not confirmed by autopsy, and Burke (*loc. cit.*) cites three such cases. The latter concludes with the assertion that the accentuation of the pulmonary second sound in pulmonary stenosis as a proof of the existence of open ductus, though possible, is supported by absolutely no facts. He believes that such accentuation is an auricular tone and is diagnostic of open foramen ovale, which was present in all three instances.

CASE IV.—*Absence of Anterior Portion of Auricular Septum.* This specimen is also from a Philadelphia Hospital case, being obtained by Dr. Coplin from a woman of thirty-two, who died of carcinoma of the uterus. There had been no clinical evidence of cardiac anomaly. The opening occupies the anterior portion of the auricular septum and is 32 mm. in diameter. Cases of partial absence of the septum are not uncommon, of the entire septum rare; instances of non-closure of the foramen ovale are exceedingly numerous. These openings, even when of large size, are usually of little or no clinical importance, appearing to exert no direct effect upon the circulation. When a large auricular opening is present in cases of mitral stenosis, blood may pass to the right auricle and give rise to jugular venous pulse in the neck. The possibility of paradoxic or crossed embolism must always be borne in mind in connection with such lesion.

In order to determine if the myocardium of malformed hearts showed histologic differences from that of normal organs, sections were made from the left ventricle in Case II. and from both left and right ventricles in Cases III. and IV. Microscopic study shows no noteworthy changes as compared with the usual histologic appearance of the normal myocardium nor is there appreciable difference between the two sides of the heart in the last two cases. The specimens, however, had been in preserving fluids for at least

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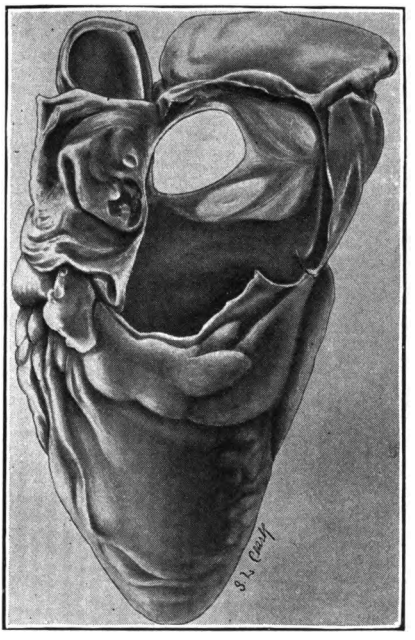
several months before this study was made and the findings cannot be regarded as conclusive.

The influence of heredity in the production of malformation of the heart appears to be positive. In my previous paper I cited several series which point to the active role of this factor. Of interest in this connection, though not exactly similar, is the case reported by Boinet.[13] A woman of twenty-three, who for three years had mitral stenosis, which followed pneumonia, died as the effect of



Case IV.

a second attack of pneumonia and aborting a six months' fetus. Autopsy on mother and fetus showed similar lesions of the mitral valve in each, the process not being so advanced in the latter, although distinct fibrous nodules were present on the leaflets. The fetus was normal in every other respect and syphilis and tuberculosis could be eliminated as causes. Boinet considers it probable that the first attack of pneumonia caused endocarditis in the mother and later this lesion was transmitted to the fetus.

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It is a pleasure to acknowledge the aid in searching the literature of this subject rendered by Max Meitzner and H. L. Hull, research students in the Jefferson Medical College.

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December 28, 1905.

Case of Cystic Left Kidney and Calculus of the Right Kidney.

BY M. H. FUSSELL, M.D.

THE patient from whom these specimens were removed was admitted to St. Timothy's Hospital August 21, 1905, with symptoms of stone in the kidney.

An *x*-ray picture showed three shadows toward the lower pole of the right kidney. No definite history was obtained except that of stone. After he had been in the hospital three days, however, he complained rather suddenly of distention of the abdomen, and we were told by his mother that he had a lump in his abdomen since his childhood; the lump was sometimes large, sometimes small, and sometimes disappeared.

On examination of the abdomen on August 25, 1905, a large mass was seen occupying the lower three-fourths of the abdominal cavity. This mass was cystic and not very painful. On the 28th, the mass had markedly increased in size, the dulness extending upward, posteriorly to the fifth rib, and laterally to the seventh rib, the mass was still painless, and there was marked fluctuation. On the 30th the

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mass extended toward the right until the right edge was midway between the umbilicus and the right side of the abdomen.

At this time the patient gave evidence of so much distress that the tumor was tapped by Dr. Wood. Soon the patient died of uremia.

An autopsy was not allowed, but both kidneys were removed through the original opening; the left was found to be a huge cystic kidney, measuring from the upper to the lower extent of the pelvis around the poles of the kidney 56 cm., the length of the kidney was 33 cm., the length of the pelvis was 20 cm., the transverse circumference with the pelvis distended was 41 cm., the longitude and circumference 72 cm.

Though the specimen was removed through a small opening the ureter was carefully examined to the bladder and no obstruction was found. The kidney consisted of a great number of cysts varying in size from that of a walnut to that of a man's fist.

Through the surface are small areas which resemble normal tissue, the capsule is greatly thickened, but somewhat easily detached. The pelvis is greatly distended, the ureter is not at all dilated, is patulous and comes off the lower part of the pelvis and is placed at the proper angle. On section of the right kidney it seems to be rather larger than normal but healthy. In the distinctly distended calices are found *eight* stones: two in the pole, both being embedded in the kidney substance, three about in the middle of the kidney, and four in the lower pole.

It seems probable from the appearance of this cyst that it is not a congenitally cystic kidney; the cysts are too large, and there is no obstruction of the ureter of congenital origin. On the other hand there is no evidence of stone, the commonest cause of postnatal hydronephrosis. It would appear from the fact that the ureter is not dilated and that there was no stone in the pelvis that obstructions might easily have occurred by a kinking of the ureter close to the pelvis of a movable kidney. It is certain from our own observation that the tumor varied in size. When the patient entered the hospital no tumor was observed, though there is no record of a careful abdominal examination. As he passed through the hands of several physicians and nurses, and had an *x*-ray taken before the tumor was observed, the mass was certainly not very large and could

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not have been the size it had attained when seen by the writer. These facts, together with the history of a tumor which had appeared and disappeared through a long series of years, make this case certainly one of intermittent hydronephrosis, due to intermittent constriction of the ureter close to the pelvis. As is well known, cysts of the kidney are either congenital or acquired. The congenital cystic kidneys are usually bilateral, have as their cause either a formation of many small cysts, or an obstruction of the pelvis or ureter by membranous divisions.

The commonest cause for acquired hydronephrosis is a stone, though a kinking of the ureter or pressure from a tumor may be the cause. Where the obstruction is in the ureter the ureter is dilated between the point of obstruction and the pelvis.

Another point of interest in the case is the multiple renal calculi in the right kidney. Eight stones is certainly a very unusual number, and the stones were faceted and had the appearance of biliary calculi.

No analysis of these stones has been made; however, the light-brown color and the faceted appearance, according to Ziegler, would mark them as probably composed of phosphate of calcium.

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